

Congenital Band about the Pelvis (A Case Report)

Chul Gyoo Park and Rong-Min Baek

Department of Plastic and Reconstructive Surgery, College of Medicine, Seoul National University, Seoul 110-741, Korea

=Abstract= Congenital band is a rare collection of fetal malformations associated with fibrous bands that appear to entangle or entrap various fetal parts in the uterus leading to deformation, malformation or disruption. Heredity, sex, and race seem to play insignificant role, and its etiologic explanation is still confusing and conjectural.

Congenital band about the pelvis is very rare and only five such documented cases are reported in the world medical literature. Now we present the sixth case of congenital band about the pelvis, treated with multiple Z-plasties, with particular consideration on the etiologic factors.

Key words: *Congenital band, Fetal malformation*

INTRODUCTION

Congenital band is a collective terminology of rare fetal malformations associated with fibrous bands that appear to entangle or entrap various parts in utero leading to deformation, malformation, or disruption. This anomaly is given many other names such as congenital constriction ring, annular band, and amniotic band syndrome, and has variety of clinical presentations.

Congenital band about the pelvis is an extremely rare entity, and only five such cases are reported in the world medical literature. A few interesting points could be extracted from the five cases. There was no sexual or racial inclination in those five cases. Three had the congenital bands above the pelvic brim, two below the pelvic brim. However, in all five cases, the bands appear to be higher posteriorly, likely following a specific somite.

None of these reports commented on the associated congenital anomalies in two cases. Schneider's case had pilonidal sinus and cleft of soft palate, Izumi and Arnold's had bands around two toes and club foot. Casaubon's case had no

associated congenital anomaly.

Only Evans' case had family history that patient's mother had a band around one toe.

Surgical correction was performed in three cases, all by Z-plasty. Two of these treated patients got pregnant and gave birth to sons without any congenital abnormality.

CASE REPORT

A 17-year-old Korean girl visited our clinic for a band-like depression around the waist (Figure 2) and short first, second, and third toes of the right foot. (Figure 1) She was born in 1966 as the younger of two siblings with no family history of congenital anomaly. The pregnancy and delivery were uneventful.

Physical examination showed depressed, constrictive, fibrous band all around the lower abdomen and back, and distal amputation of first, second, and third toes of the right foot, which proved to be absence of the distal phalanges by X-ray evaluation. Otherwise there was no remarkable abnormal finding.

In 1983, multiple Z-plasties after excision of the band were performed for the release of the constriction ring about the pelvis (Figure 3). The biopsy specimen showed the epidermis to be normal. The dermis was thick, composed of collagen bundles and elastic fibers, nearly typical

ing the subcutaneous tissue.

The adnexal structures were normal. (Figure 5, 6) There was no more constriction after Z-plasties.

DISCUSSION

Of five reported cases of congenital band about the pelvis, Izumi and Arnold's case is similar to ours. However, generalization of all six cases (including our case) seems to be difficult; Sex, race, and heredity appear to play a very limited role. The congenital band about the pelvis does not seem to offer a real functional constriction, rather it seems to be a cosmetic problem because it allowed normal pregnancies with or without Z-plasty in two cases. The location of the bands is almost uniform, along a specific somite, higher posteriorly. Four of six cases were treated by Z-plasty. Since Stevenson's suggestion in 1946, Z-plasty has been the most common method for the release of congenital band.

There are extreme controversies on the etiology of congenital band. In 1930, Streeter proposed his theory of defective germ plasm. After studying 16 preserved laboratory specimens of amniotic band syndrome he concluded that congenital band were due to developmental defects of the germ disk and imperfect histogenesis in the developing tissue lead to a localized sclerosis. Schneider supported Streeter's view with his microscopic findings of his case, which resemble the discription of Streeter of "the these...strands...proved to be the hyalinized product of defective tissue." Keith suggested that Streeter's fetal dysplasia was due to early placental circulatory disturbance. However, Streeter's view of intrinsic etiology failed to explain the inconsistencies of asymmetry, sporadic occurrence, and the absence of related internal anomalies.

An alternative theory for the etiology of congenital band was published in 1965 by Torpin. After the study of the placenta of the patient with congenital band he concluded mesodermic band (amniotic and chorionic fibrous strings) formed following rupture of the amnion could encircle fetal parts and similarly produce the characteristic constriction ring. In 1981, Mayou and Fenton reported in their study of six cases of facial cleft that an amniotic string was found lying deep in the bilateral oblique facial cleft. In

1982, Opgrande suggested their case of congenital band to be the result of extrinsic pressure produced by the fibrous band that was thought to be amnion on the microscopic examination. In 1975, Kino concluded in his study using rats that the constriction ring was the result of excessive contraction of the uterine muscle and hemorrhage from the marginal blood sinuses, and suggested the malformations were not hereditary, but are probably produced by prenatal environmental factors.

Although the majority of recent papers support Torpin's view that congenital band results from extraamniotic, intrachorionic entanglement with mesodermic strands, and that theory seems best able to explain the broad spectrum of clinical features, the etiologic explanations remain confusing and conjectural, and need to be further clarified.

We think a rather specific location of congenital band about the pelvis favors the Streeter's view of intrinsic etiology.

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= 국문초록 =

체간에 발생한 선천성 수축반흔환

서울대학교 의과대학 성형외과학교실

박철규 · 백룡민

선천성 수축반흔환은 섬유성 띠(band)가 자궁내 태아신체의 일부를 엮든지 감아서 변형 또는 절단을 유발하는 태아기형의 일종으로 이제까지 annular band, congenital constriction band syndrome, congenital ring constriction, amniotic band syndrome, amniogenic band 등의 여러 병명으로 불려져 왔다.

임상적으로는 많은 경우 다른 여러가지 기형을 동반할 수 있으며, 주로 사지에 발생하고, 두안면기형이나 내장 기형도 흔히 동반하는 것으로 알려졌으며, 이중 체간부에 생기는 수축반흔환은 극히 희귀하여 현재까지 문헌상으로는 전세계적으로 5례만이 보고되었을 뿐이다.

저자는 요부 주위에 생긴 희귀한 체간 선천성 수축반흔환 1례를 Z-성형술로 치료하고 좋은 결과를 얻었기에 문헌고찰과 함께 보고하는 바이다.

LEGEND FOR FIGURES

- Fig. 1. Distal amputation of right first, second, and third toes.
- Fig. 2. Congenital band about the pelvis. Note the band is higher posteriorly.
- Fig. 3. Postoperative view after multiple Z-plasties (abdomen).
- Fig. 4. Postoperative view after multiple Z-plasties (back).
- Fig. 5, 6. Thick dermis with collagen bundles extending below eccrine glands.

